Anesthetic management of a patient with Bartter's syndrome undergoing bilateral sagittal split osteotomy

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ABSTRACT

Bartter's syndrome is an unusual (estimated incidence is 1.2 per million people) but important congenital form of secondary hyperaldosteronism; due to abnormalities in renal handling of electrolytes. It is associated with hypertrophy and hyperplasia of the juxtaglomerular cells, normal blood pressure, and hypokalemic alkalosis withoutedema. We present a 22-year-old woman with Bartter's syndrome underwent bilateral sagittal split osteotomy to correct mandibular prognathic. The anesthetic management of Bartter's syndrome should be relevant to the pathophysiology of the syndrome. Therefore, it should be directed toward maintaining cardiovascular stability, control of associated fluid, electrolyte and acid–base derangements, and the prevention of renal damage.

Key words: *Bartter's syndrome, bilateral sagittal split osteotomy, orthognathic surgery*

INTRODUCTION

Bartter's syndrome is a heterogenous entity with at least 2 subsets: Hypokalemic alkalosis with hypercalciuria (true Bartter's syndrome) and hypokalemic alkalosis with hypercalciuria (Gitelman's syndrome). It can also present in utero (antenatal Bartter's syndrome) with resulting prematurity or polyhydramnios.[1] The pathogenesis of Bartter's syndrome is obscure. The primary (autosomal recessive) defect lies in the active chloride reabsorption in the loop of Henle.[1] There is loss of excessive amounts of sodium and potassium in the urine, which leads to hypovolemia and secondary hyperaldosteronism.[2] True Bartter patients usually present above 5 years with signs of vascular volume depletion, polyuria, and polydipsia, whereas Gitelman's syndrome patients typically present at older ages without overt hypovolemia as failure to thrive.[3] Other features include the following: Impaired urinary concentrating ability and hyperactive renin–angiotensin system (plasma renin increased, lack of effect of angiotensin on blood pressure, renal potassium wasting, increased renal prostaglandin production, and occasionally hypomagnesemia.[2]

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CASE REPORT

Our patient is a 22-year-old Saudi girl, had mandibular prognathism (class III malocclusion with 5 mm discrepancy), and stunted stature, weighing 41 kg, a known case of Bartter's syndrome since childhood and currently complicated by nephrosclerosis. She was scheduled for bilateral sagittal split mandibular osteotomy (BSSO) under general anesthesia. After 2 years of orthodontic preparation, and after discussing all the details of the surgery with its suspected complications with the patient and her family. The patient was insisting for the surgery in spite of her medically challenging condition. In her family the disease is inherited as an autosomal recessive trait (her brother suffers the same condition). She has a previous history of receiving growth hormone at the age of puberty as well as a history of receiving indomethacin; however, discontinued by the nephrologists.

The patient was seen in the preanesthesia clinics 2 weeks before the predetermined date of surgery. Her current medications were potassium chloride 600 mg and calcium carbonate 600 mg taken 2 times daily; spironolactone 100 mg, allopurinol 100 mg, and calciterol 0.5 mg taken once daily. The investigations showed a hypokalemic, hypochloremic, metabolic alkalosis Na^+135, K^+2.3, Cl^-92, and a $HCO3^-$35 mmol/L. The patient's oral potassium chloride intake was increased to 600 mg 3 times daily.

On admission to the hospital (2 days preoperatively), the patient's physical examination was normal, and blood pressure was 110/75 mmHg. The electrocardiogram demonstrated only nonspecific ST-T changes. Laboratory data indicated that the electrolyte derangement was corrected; serum sodium, potassium, and chloride were 147, 4.4 mmol/L, and 107 mmol/L, respectively [Table 1]. Calcium (2.67 mmol/L), corrected calcium (2.7 mmol/l mol/L) and magnesium (0.8 mmol/L) levels were all within normal. Urea and creatinine were slightly elevated indicating underlying renal disease (8 mmol/L and 117 μmol/L, respectively). The patient was neither anemic (hemoglobin 124 g/L) nor had high hematocrit (36%). Laboratory results obtained in a day before or immediately before the surgery were within normal ranges; except for blood urea and creatinine that was still increasing and the nephrologist was consulted [Table 1]. It was decided to proceed to the surgery without any further correction.

Patient's medications were continued till the day of surgery. Clindamycin 600 mg and dexamethazone 6 mg intravenously were administered at the night before the surgery as well as on the induction of anesthesia as recommended by the surgeons. The patient was premedicated 60 min preoperatively with lorazepam, 1 mg, ranitidine 150 mg, and metoclopramide 10 mg orally.

Before induction of anesthesia, percutaneous radial arterial was inserted. The insertion of central venous pressure catheters was discussed and decided not to be inserted. Intravenous fluid replacement for the hours of fasting (D5 I/2N saline) was completed before starting anesthesia.

The anesthesia was induced intravenously by high-dose fentanyl 5 μg/kg, propofol increments (total of 40 mg), and cisatracurium 6 mg. After nasal preparation with vasoconstrictor; cuffed endotracheal tube size 6.5 mm was inserted. Mechanical ventilation was initiated bearing in mind to avoid hyperventilation. With tidal volume of 400 mL and respiratory rate of 10, end-tidal CO_2 was maintained between 34 and 37 mmHg. Besides the standard monitoring, urine output, neuromuscular assessment, and bispectral index were also included.

Arterial blood pressure before induction of anesthesia was 120/70 mmHg and heart rate 82 beats/min. Systolic blood pressure never exceeded 150 mmHg or decreased below 100 mmHg. However, heart rate increased over 130 beats/min immediately after infiltration of local anesthetic (xylocaine 2% with adrenaline 1/100,000) into the surgical field. IV labetalol 15 mg (increments of 5 mg) was given until the tachycardia was controlled. Two measurements of serum electrolyte and blood gas levels were done intraoperatively and there were no significant changes [Table 1].

Surgical procedure was as follows: BSSO cuts were done intraorally following the technique described by Nooh and Abdullah (2007),[4] then a 7 mm straight forward setback of

Table 1: Biochemical data

Parameters (Normal range)	Preoperative			Intraoperative		Postoperative			
	Admission	1 Day (Pre)	0900 hours	1100 hours	1200 hours	Day 1		Day 2	
						1400 hours	2200 hours	0600 hours	1400 hours
Sodium (135–145 m mol/L)	147	143	140	124	135	140	135	142	140
Potassium (3.5–5 mmol/L)	4.4	4	3.8	3.8	4.2	4	3.0	3.6	4
Chloride (95–105 mmol/L)	107	104	98	101	100	101	90	104	101
Magnesium (0.7–1.1 mmol/L)	0.8					0.6			
Urea (2.5–6.6 mmol/L)	8	10.1	10.3	9	9.3	9.1	8.5	8.9	9.1
Creatinine (53–106 μmol/L)	117	151	140	131	131	131	140	138	131
Calcium (2.1–2.55 mmol/L)	2.67	2.89	2.92			2.6	2.06	1.93	2.6
Calcium corrected (3.5–5 mmol/L)	2.7	2.9	2.9			2.5	2.2	2.1	2.5
pH (7.36–7.44)				7.45	7.34	7.38	7.49	7.45	7.45
Bicarbonate (22–26 mmol/L)				24.6	24.1	24	28.3	25.7	23.9
PaO_2*(83–112) mm Hg				202	203	141	131	92	95
$PaCO_2$ (33–45) mm Hg				34	36	41	35	35	35
Hemoglobin (Female: 14±2 g/dL)	12.4	12.4	12.5	11.0	10.8	11.6	10.8	10.7	10.6
Hematocrit (Female: 41±5%)	36	36	34	31	32	35	34	34	34

*PaO2=according to FIO2, FIO2=0.5 intraoperatively, oxygen mask and room air in day 1 and day 2 postoperatively, respectively

the mandible was done and fixed using one 2 mm titanium plate and 4 screws (2 screws of 5 mm length anterior to the bone cut, and 2 screws of 7 mm length posterior to the bone cut) at each side.

The surgical time was 90 min, bleeding was minimal; vasopressors to maintain the blood pressure were not required. Intraoperative fluid balance was as such (Intake = 1000 mL Lactate Ringer solution and 500 mL D5 I/2N saline and the output; urine output (UOP) = 700 mL/3 h). The patient tolerated the procedure very well.

The patient was transferred to the surgical intensive care unit (SICU). Intravenous fluid D5 I/2N saline 80 mL/h together with KCl 20 meq/L every 6 h was commenced till the patient resumed oral intake. Magnesium level dropped to 0.6 mmol/L. $MgSO_4$ 2 g was given as IV infusion over 2 h twice daily. Ca gluconate 1 g was ordered if serum calcium level fell below 2 mmol/L and only one dose was administered. Electrolytes were measured on an 8-hourly basis. UOP did not surpass 100 mL/kg/day in the postoperative period. However, at the night of surgery the patient showed hypokalemic, hypochloremic, and metabolic alkalosis [Table 1]. Na^+135, K^+3.0, Cl^-90, HCO_3^-28.3 mmol and a pH 7.49. Intravenous fluids were changed to normal saline and intravenous potassium chloride replacement increased to 30 meq/6 h, next day the oral potassium chloride was gradually introduced instead of the intravenous replacement. Eventually, the patient was discharged 3 days postoperatively on the same treatment she had received preoperatively.

DISCUSSION

Anesthetic management of a patient with Bartter's syndrome presents many challenges to anesthesiologist. In the light of pathophysiology of Bartter's syndrome, cardiovascular instability, perioperative electrolyte and acid–base disturbances, and renal dysfunction are the most concerned.

The stability of cardiovascular system is important in such patients.[2] Major hemodynamic problems were not encountered in patients with Bartter's syndrome.[5-7] They are normotensive although they have tendency to dehydration. Sodium chloride wasting and long time treatment with potassium-sparing diuretics, spironolactone, are the common causes of expected hypovolemia. In addition, however, not applied in our case, patients may be on myocardial beta-adrenergic blockade, propranolol; there may be no chronotropic response in the hypovolemic patient or may be on angiotensin-converting enzyme inhibitors and significant hypotension may follow when there is fluid loss during anesthesia.[2]

In the present case, percutaneous radial arterial catheter was inserted preoperatively and urinary output was monitored using a Foley catheter, however, we thought unnecessary to insert central venous pressure catheter. Reading of the central venous pressure may be interrupted by the surgical procedure and positioning of the patients. Major bleeding or fluid derangements is unusual in such surgeries. In addition, the patient did not encounter any incidence of brisk diuresis or show any sign of dehydration in the preoperative period. To eliminate any cause that lead to hypovolemia, intravenous fluid replacement for the hours of fasting was compensated before starting anesthesia. Hematocrit was not elevated before surgery [Table 1]. A high hematocrit may reflect an absolute increase in the number of erythrocytes, or a decrease in plasma volume.[8] The reasons for absolute increase in the number of erythrocytes are not present in the case. Therefore, dehydration (a decrease in plasma volume) would have been reflected by increased hematocrit values. Few case reports have described management for patients with Bartter's syndrome under general anesthesia. Kannan *et al.* described the anesthetic management of a child with Bartter's syndrome by caudal epidural analgesia for elective orchidopexy without measuring central venous pressure.[7] Roelofse and Van der Westhuijzen managed a patient with Bartter's syndrome,' who required orthognathic surgery in their case report.[9] Because of the possibility of hypovolemia, the placement of a central venous pressure was thought to be mandatory. However, the central venous pressure was easily kept between 4 and 10 cm H_2O. Vetrugno *et al.* reported a case described a patient with Bratter's syndrome who underwent cardiac anesthesia without any major complication. In the authors' experience, invasive cardiovascular monitoring including pulmonary artery catheter is recommended in measuring the volume depletion, and mannitol should not be used in the priming solution.[10]

Hemodynamic parameters were stable all through the surgery except with that incidence of sinus tachycardia after local anesthetics with adrenaline infiltration of the surgical wound. This was not expected in Bartter's syndrome patients with decreased pressor responses. Frederic Crosby Bartter when first described the syndrome in 1962 based on 2 patients, reported decreased pressor responsiveness to angiotensin II infusion.[11] Nishikawa and Dohi investigated the baroreflex function in a 40-year-old woman with Bartter's syndrome.[12] They concluded that unstable baroreceptor responses exist in patients with this syndrome.

Patients with Bartter syndrome have markedly raised basal renin and high levels of angiotensin; which can lead to nonresponsiveness of their blood vessels to angiotensin. This could be the reason why they do not develop high blood pressure. Nonetheless, it has now been demonstrated

that patients with Bartter's syndrome will show a normal response to vasopressor agents once their volume is restored to normal.[13] Presumably, this was the case; our patient had a response to the absorbed adrenaline when the local anesthetic was infiltrated.

Judicious attention of electrolyte and acid–base status are of utmost importance not only in the intraoperative period but also during the entire perioperative period. It should individually be titrated in accordance to the patient's needs. This patient in the pre-anesthesia clinic showed hypokalemic, hypochloremic, and metabolic alkalosis; it was decided for her to increase her oral potassium chloride intake 600 mg 3 times a day. This was capable to adjust her biochemical results before the surgery. This was extended in the intraoperative period and early in the postoperative period. Oral supplementation therapy was proved to be almost totally ineffective by itself, since administered potassium is lost through the kidney in a short period of time.[1] Therefore, the concomitant use of potassium-sparing agents, such as spironolactone, as it offers an effective control of hypokalaemia may seem logical.[1]

At the night of surgery the patient developed hypokalemic, hypochloremic, and metabolic alkalosis [Table 1]. Intravenous normal saline, instead of D5 I/2N was commenced and intravenous potassium chloride replacement increased to 30 meq/6 h, next day the oral potassium chloride was gradually introduced instead of the intravenous replacement judged with laboratory results till potassium level reached its normal level. Potassium wasting can be exaggerated by magnesium deficiency.[1] Therefore, $MgSO_4$ 2 gm over 2 h twice daily was added to the postoperative therapeutic regimen and one dose of calcium gluconate when serum calcium level falls below 2 mmol/L.

Considerations in the anesthetic technique should be in the view of the pathophysiologic and biochemical changes in patients with Bartter's syndrome, however, should individually be titrated tailored in accordance to the patient's needs. We induced anesthesia in a narcotic-based pattern to reduce the amount of intravenous anesthetics, which can cause hypotension if the patient was hypovolemic. Intravenous fluid replacement was also administered immediately before surgery. Since patients with Bartter's syndrome have low serum potassium, hyperventilation should be avoided, as hypocapnia increases blood pH and lowers serum potassium.[14] The effect of neuromuscular-blocking drugs during anesthesia should also be monitored with a nerve stimulator in patients with hypokalemia. The renal function of patients with Bartter's syndrome may affect the choice of the anesthetic agents. Elimination of volatile anesthetic agents is independent of renal function. Isoflurane was used before in those patients and we used sevoflurane in this report. However, there have been isolated reports of possible fluoride-induced nephrotoxicity with enflurane, which should probably be avoided.[15]

Although anesthesia management in patients with Bartter's syndrome requires judicious attention with respect to maintaining of cardiovascular stability, control of plasma potassium level and the prevention of renal damage, it should be individually tailored in accordance to the surgical procedure and patient's clinical condition.

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